Management of Patients with Orofacial Clefts

CHAPTER

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CHAPTER OUTLINE

A cleft is a congenital abnormal space or gap in the upper lip, alveolus, or palate. The colloquial term for this condition is *harelip.* The use of this term should be discouraged, because it carries demeaning connotations of inferiority. The more appropriate; terms are *cleft Sip, cleft palate,* or *cleft lip and palate.*

Clefts of the lip and palate are the most common serious congenital anomalies to affect the orofacial region, Their initial appearance may be grotesque. Because they are deformities that can be seen, felt, and heard, they constitute a serious affliction to those who have them. Because of their location, they are deformities that involve the dental specialties throughout their protracted course of treatment. The general dentist will become involved in managing these patients' special dental needs, because they may have partial anodontia and supernumerary teeth. Malocdusion is usually present, and orthodontic therapy with or without corrective jaw surgery is frequently indicated.

The occurrence of a cleft deformity is a source of considerable shock to the parents of an afflicted baby, and the most appropriate approach to the parents is one of informed explanation and reassurance. They should be told that the defects are correctable and need not adversely affect the child's future. However, they should be prepared for a protracted course of therapy to correct the cleft deformities and to allow the individual to function with them.

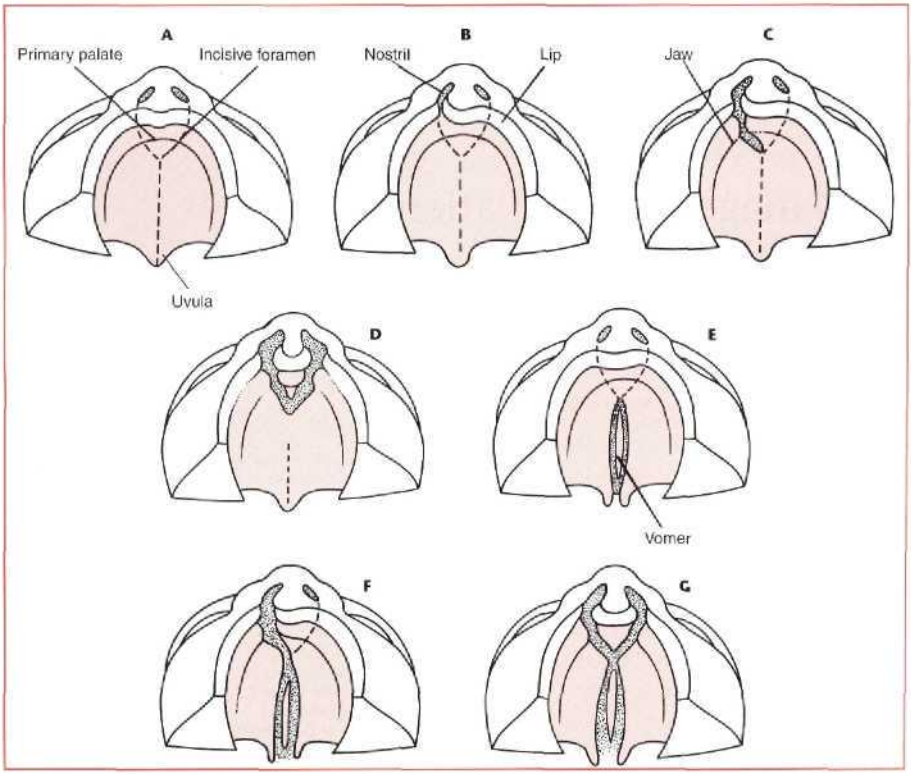


FIG. 27-1 Ventral view of palate, lip, and nose showing variability of cleft lip and palate deformity. A, Normal. B, Unilateral cleft lip extending into nose. C, Unilateral deft invoking lip and alveofus, extending to incisive foramen. D, Bilateral cleft involving lip and alveolus, E, Isolated cleft palate. F, Cleft palate combined with unilateral cleft of alveolus and lip. G, Bilateral complete cleft of lip and palate- *(From Longman I:* Medical embryology, *ed 3, Baltimore, T97S, Williams & Witkins.)*

The problems encountered in rehabilitation of palicnts with cleft deformities are unique. Treatment must address patient appearance, speech, hearing, mastication, and deglutitiun. A team manages most children currently affected with orofacial clefts,

Cleft teams are found in most cities of at least moderate size. These teams commonly comprise a general or pediatric dentist, an orthodontist, a prosthodontist, an oral and maxillofacial surgeon or a plastic surgeon, an audiologist, an otorhinolaryngologist, a pediatrician, a speech pathologist, a psychologist 01 psychiatrist, and a social worker. The number of specialists required reflects the number and complexity of the problems faced by individuals with orofadal clefts.

The occurrence of oral defts in the Uniled States has been estimated as 1 in 700 births.[1] Clefts exhibit interesting racial predilections, occurring less frequently in blacks but more so in Asians. Boys are affected by orofacial clefts more often than girls, by a ratio of *3:2.* Cleft lip and palate (together) occurs about twice as often in boys as in girls, whereas isolated clefts of the palate (without cleft lip) occur slightly more often in girls.

Oral clefts commonly affect the lip, alveolar ridge, and hard and soft palates. Three fourths are unilateral deformities; one fourth are bilateral. The left side is involved more frequently than the right when the defect is unilateral. The cleft may be incomplete, that is, it may not extend the entire distance from lip to soft palate. Cleft lip may occur without clefting of the palate, and isolated cleft palate may occur without clefting of the lip (fig. 27-1), A useful classification divides the anatomy into primary and secondary palates. The primary palate involves

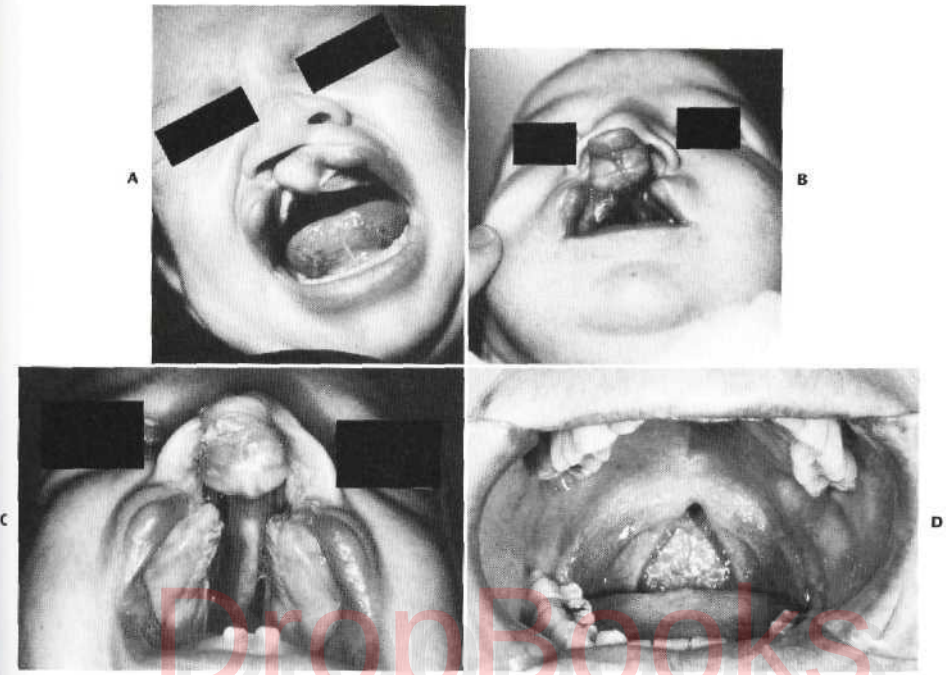


FIG. 27-2 Photographs of various types of dell deformities, Nasal deformities are also apparent. A, Unilateral compleLe cleft of lip and palate. B, Bilateral complete cleft lip and palate. C, Palatal view of bilateral complete cleft lip and palate. The nasal septum is unattached to either palatal shelf, D, isolated cleft of soft palate.

those structures anterior to the incisive foramen—the lip and alveolus; the secondary palate consists of those structures posterior to the incisive foramen—the hard and soft palates/ Thus an individual may have clefting of the primary palate, the secondary palate, or both (Fig. 27-2).

Clefts of the lip may range from a minute notch on the edge of the vermilion horder to a wide cleft that extends into the nasal cavity and thus divides the nasal floor. Clefts of the soft palate may also show wide variations from a bifid uvula to a wide inoperable cleft. The bifid uvula is the most minor form of cleft palate. In which only the uvula is clefted. Submucosal clefts of the soft palate, are occasionally seen. These clefts are also called *occult* clefts, because they arc not readily seen on cursory examination. The defect in such a cleft is a lack of continuity in the musculature of the soft palate. However, the nasal and oral mucosa is continuous and covers the muscular defect. To diagnose such a defect, the dentist inspects the soft palate while the patient says *"ah,"* This action lifts the soft palate, and in individuals with subrtiucosal palatal clefts, a furrow in the midline is seen where the muscular discontinuity is present. The dentist can also palpate the posterior aspect of the bard palate to detect the absence of the posterior nasal spine, which is characteristically absent in submucosal clefts. If a patient shows hypernasal speech without an obvious soft palatal cleft, the dentist should suspect a submucosal cleft of the soft palate.

EMBRYOLOGY

To understand the causes of oral clefts, a review of nose, lip, and palate embryology is necessary. The entire process takes place between the fifth and tenth weeks of fetal life.[3]

During the fifth week, two fast-growing ridges, the *lateral* and *medial nasal swellings*, surround the nasal vestige (Fig. 27-3), The lateral swellings will form the alae of the nose; the medial swellings will give rise to four areas: (1) the middle portion of the nose, *(2)* the middle portion of the upper lip, (3) the middle portion of the maxilla, and (4) the entire *primary palate*. Simultaneously the maxillary swellings will approach the medial and lateral nasal swellings bul remain separated from them by well-marked grooves.

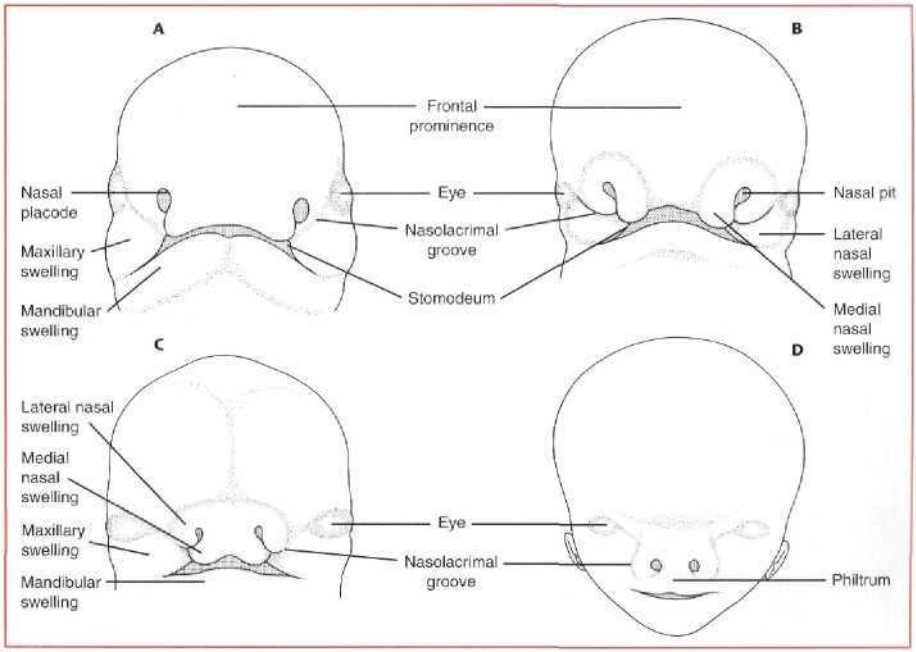


FIG. 27-3 Frontal aspect of face. A, Five-week-old embryo. B, Six-week-old embryo, Nasal swellings art gradually separated from maxillary swelling by deep furrows. At no time during normal development doe; this tissue break down. C, Seven-week-old embryo. D, Ten-week-old embryo. Maxillary swellings gradually merge with nasal folds, and furrows are filled with mesenchyme. *(From Longman j:* Medical embryology, *ed 3, Baltimore, 1 975, Williams £r Wilkins.)*

During the next 2 weeks, the appearance of the face changes considerably. The maxillary swellings continue *to* grow in a medial direction and compress the medial nasal swellings toward the midline. Subsequently these swellings simultaneously merge with each other and with the maxillary swellings laterally. Hence the upper lip is formed by the two medial nasal swellings and the two maxillary swellings.

The two medial swellings merge not only at the surface but also at the deeper level. The structures formed by the two merged swellings are known together as the *intermaxillary segament* (Fig. 27-4)which is comprised of three components: (1) a labial component, which forms the philtrum of the upper lip; (2) an upper jaw component, which carries the- four incisor teeth; and (3) a palatal component, which forms the triangular primary palate. Above, the intermaxillary segment is continuous with the nasal septum, which is formed by the frontal prominence.

Two shelflike outgrowths from the maxillary .swellings form the secondary palate. These *palatine shelves* appear in the sixth week of development and are directed obliquely downward on either side of the tongue. In the seventh week, however, the palatine shelves ascend to attain a horizontal position above the tongue and fuse with each other, thereby forming the *secondary palate.* Anteriorly the shelves fuse with the triangular primary palate, and the incisive foramen is formed at this junction. At the same time, the nasal septum grows down and joins the superior surface of the newly formed palate. The palatine shelves fuse with each other and with the primary palate between the seventh and tenth weeks of development.

Clefts of the primary palate result from a failure of me.soderm to penetrate into the grooves between the medial nasal and maxillary processes, which prohibits their merging with one another. Clefts of the secondary palate are caused by a failure of the palatine shelves to fuse with one another. The causes for this arc? speculative and include failure of the tongue to descend into the oral cavity,

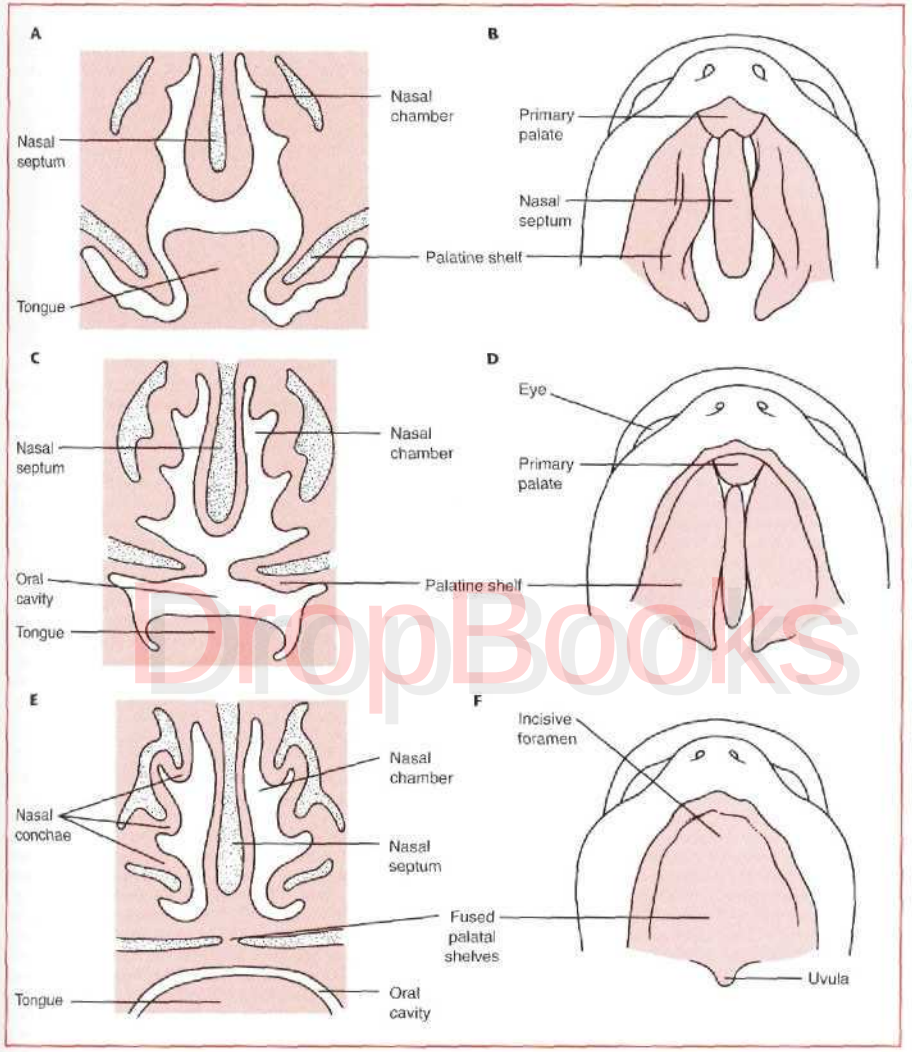


FIG. 27-4 A, Frontal section through head o(6^{1} /a-weeK-old embryo. Palatine shelves are located fn vertical position on each side of tongue, B, Ventral view of same, Note clefls between primary triangular palate and palatine ihelves, which arc itill in vertical position. C, Frontal section through head of 7V2-week-old embryo, Tongue *has* rtiovrd downward, and palatine shelves have reached horizontal position. O, Ventral view of same. Shelves are in horizontal position, E, Frontal section through head of 10-week-old embryo. Two paiatine shelves have fused with each other and with nasal septum. F, Ventral view of 5ame. *(From Longman):* Medical embryofogy, *ed _?, Baltimore, 1* 9/5, *Williams & WHkins.)*

CAUSATIVE FACTORS

The causes of facial clefting have been extensively investigated. The exact cause of cleftiny is unknown in most cases. For most cleft conditions, no single factor can be identified as the cause. However, it is important to distinguish between *isolated clefts* (in which the patient has no other related health problem) and *clefts associated with other birth disorders or syndromes.* A syndrome is a set of physical, developmental, and sometimes behavioral traits that occur together. Clefts have been identified as a feature in over 300 syndromes, most of which are rare.[1] Syndromes account for approximately 15% of the total number of cases of cleft lip and cleft palate but nearly 50% of cases of isolated cleft palate. Medical geneticists are usually asked to consult with the family of children born with syndromes to identify the specific syndrome and to provide information to the parents about the likelihood of another child being affected.

For nonsyndromic clefts, it was initially thought that heredity played a significant role in the causation. However, studies have been able to implicate genetics in only 20% to 30% of cleft lip or palate patients. Even in those individuals whose genetic backgrounds may verify familial tendencies for facial clefting, the mode of inheritance is not completely understood. It is not *a* simple case of mendelian dominant or recessive inheritance but is multigenetic. The majority of nonsyndromic clefts appear to be caused by an interaction between the individual's genes (i.e., genetic predisposition) and certain factors In the environment that may or may not be specifically identified.

Environmental factors seem to play a contributory role at the critical time of embryologic development when the lip and palatal halves are fusing. A host of environmental factors have been shown in experimental animals to result in clefting. Nutritional deficiencies, radiation, several drugs, hypoxia, viruses and vitamin excesses or deficiencies can produce clefting in certain situations.

The risk for having another child with a cleft is based upon a number of factors that are often unique in a particular family- These include the number of family members with clefts, how closely they are related, the race and sex of the affected individuals, and the type of cleft each person has. After a syndrome or complex disorder is excluded, recurrence risk counseling for a cleft can be offered to families. No genetic test can determine a person's individual chance of having a child with a cleft.

Every parent has approximately a 1 in 700 risk of having a child with a cleft,[1] Once parents have a child with a cleft, the risk that the next child (and each succeeding child) will be affected is 2% to 5% (i.e., 2 to 5 chances in lOtl).[1] If more than one person in the immediate family has a cleft, the risk rises to 10% to 12% (i.e., roughly 1 chance in 10). A parent who has a cleft has a 2% to 5% chance that his or her child will have a cleft. If the parent with a cleft also has a close relative with a cleft, the risk increases to 10% to 12% for their child being born with a cleft. The unaffected siblings of a child with a cleft have an increased risk of having a child with a cleft (1% or 1 in 100, compared with 1 in 700 when no history of cleft exists). If a syndrome is involved, the risk for recurrence within a family can be as high as 50%.' Genetic counselors may be consulted for parents of children with clefts or for people with clefts who would like to obtain more information on the relative risks for their offspring.

In summary orofaclal clefts are produced by incompletely understood mechanisms, both genetic and environmental. With lack of complete knowledge of the causes, effective preventive measures, other than good prenatal practices (e.g., avoiding medications that are not absolutely necessary), are not available to prevent this deformity from developing.

PROBLEMS OF CLEFT-AFFLICTED INDIVIDUALS

Dental Problems

A cleft of the alveolus can often affect the development of the primary and permanent teeth and the jaw itself.[4] The most common problems may be related to congenital absence of teeth and, ironically, supernumerary teeth (Fig. 27-5). I he cleft usually extends between the lateral incisor and canine area. These teeth, because of their proximity to the cleft, may be absent; when present, they may be severely displaced so that eruption into the deft margin is common. These teeth may also be morphologically deformed or hypomineralized. Supernumerary teeth occur frequently, especially around the cleft margin[1];. These teeth usually must be removed at some point during the child's development. However, they may be retained if they can furnish any useful function in the patient's overall dental rehabilitation. Frequently, supernumerary teeth of the permanent dentition arc left until 2 to 3 months before alveolar cleft bone grafting, because these teeth, although nonfunctional, maintain the surrounding alveolar bone. If extracted earlier, this bone may resorb, making the alveolar cleft larger.

Malocclusion

Individuals affected with cleft deformities, especially those of the palate, show skeletal discrepancies between the size, shape, and position of their jaws. Class 111 malocclusion, seen in most cases, is caused by many factors. A common finding is mandibular prognathism, which is frequently relative and is caused more by the retmsion of the maxilla than by protrusion of the mandible (i.e., pseudoprognathism) (Fig. 27-6). Missing or extra teeth may partially contribute to the malocclusion. However retardation of maxillary growth is the factor most responsible fur the malocclusion. Generally the operative trauma of the cleft closure and the resultant fibrosis (i.e., scar contracture) severely limit the amount of maxillary growth and development that can take place. The maxilla may be deficient in all three planes of space, with rctrusion, constriction, and vertical underdevclopment common. Unilateral palatal clefts show collapse of the cleft side of the maxilla [i.e., the lesser segment) toward the center of the palate, which produces a narrow dental arch. Bilateral palatal clefts show collapse of all three segments or may have constriction of the posterior segments and protrusion of the anterior segment.

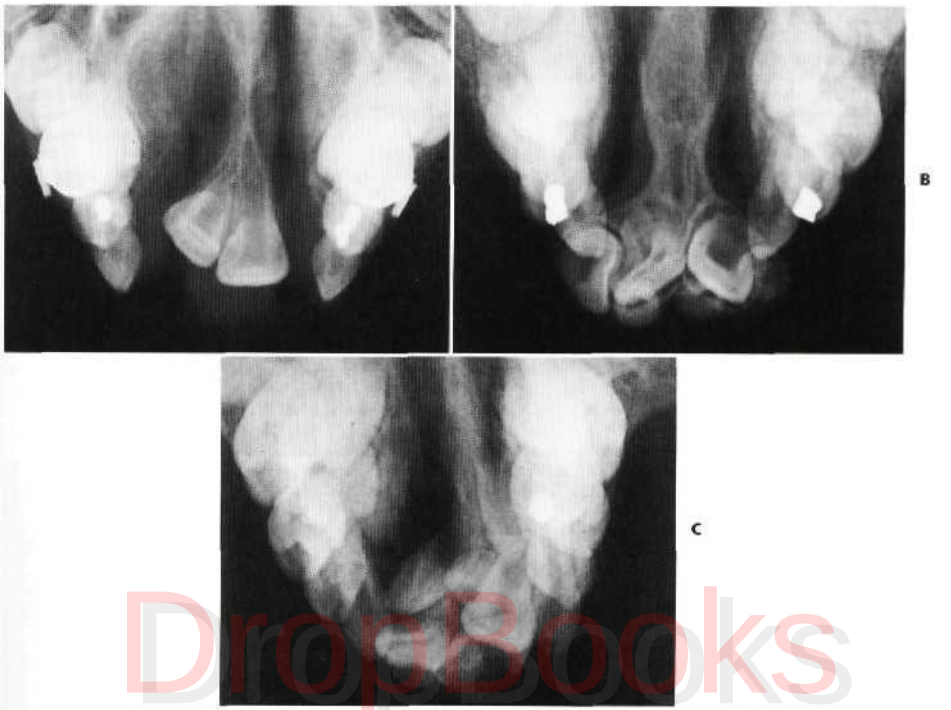


FIG. 27-5 Ocdusal radiographs from individuals wilh various types ot cleft deformities, A, Bilateral complete cleft of alveolus and paldle. Note absence of permanent lateral incisors. B, Bilateral complete deft of alveolus and palate. Note absence of permanent lateral incisor on patient's left side. C, Unilateral compfete cleft of alveolus and palate. Note .supernumerary teeth.

Orthodontic treatment may be necessary throughout the individual's childhood and adolescent years. Space maintenance and control is instituted during childhood. Appliances to maintain or incerease the width of the dental arch are frequently used. This treatment is usually begun with the eruption of the first maxillary permanent molars.

Comprehensive orthodontic care is deferred until later, when most of the permanent teeth have erupted. Consideration for orthognathic surgical intervention for correction of skeletal discrepancies and occlusal disharmonies is frequently necessary at this time.

Nasal Deformity

Deformity of normal nasal architecture is commonly seen in individuals with cleft lips (see Fig. 27-2). If the cleft extends into the floor of the nose, the alar cartilage on that side is flared and the columella of the nose is pulled toward the noncleft side. A lack of underlying bony support to the hase of the nose compounds the problem.

Surgical correction of nasal deformities should usually be deferred until all clefts and associated problems have been corrected, because correction of the alveolar cleft defect and the maxillary skeletal retrusion will alter the osseous foundation of the nose, improved changes in the nasal form will therefore result from these osseous procedures. Thus nasal revision may be the last corrective surgical procedure the cleft-afflicted individual will undergo.

Feeding

Babies with cleft palates can swallow normally once tht material being fed reaches the hypopharynx but have extreme difficulty producing the necessary negative pressure in their mouth to allow sucking either breast or bottle milk. When a nipple is placed in the baby's mouth, he or she starts to suck just like any other newborn, because the

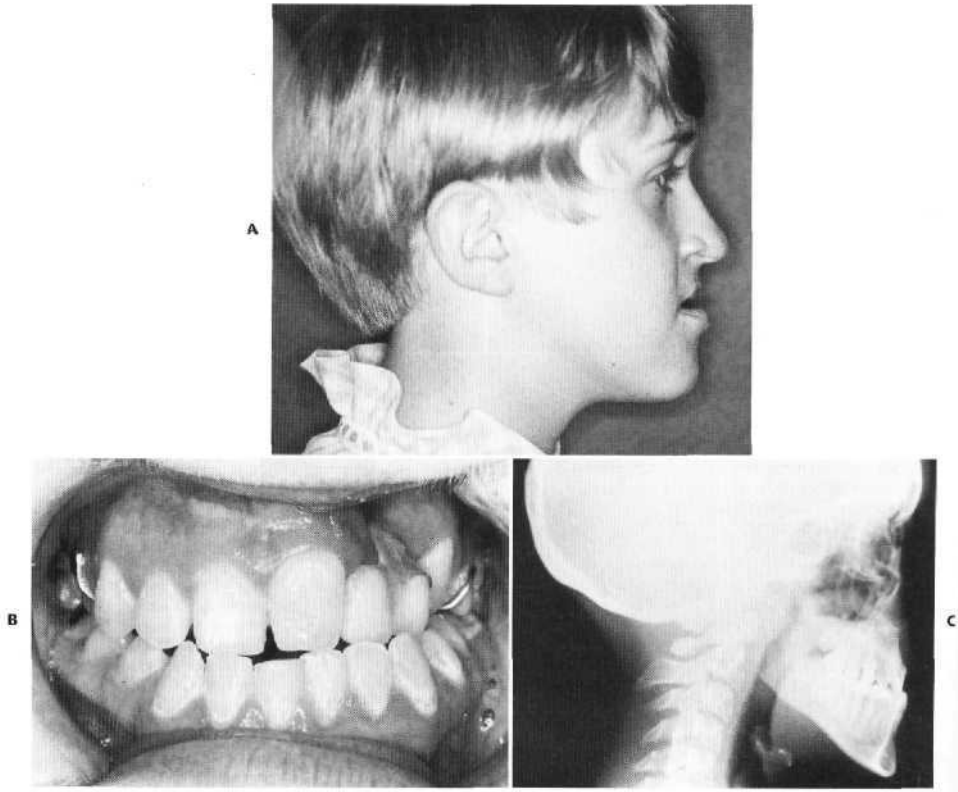


FIG. 27-6 A, *facial* profile of typical cfelt patient. Note **pseudoprognathic** appearance of mandibJe. B, Oedusal relationship ol palient showing Angle's Cass III relationship with anterior crossbite. C, **Lateral** cephaJagram showing maxillary skeletal sacjfttal deficiency contributing to Cass Hf occlusal relationship.

sucking and swallowing reflexes art normal. However, the musculature *is* undeveloped or not property oriented to allow the sucking to be effective. This problem is easily overcome through the use of specially designed nipples that are elongated and extend further info the baby's mouth. The opening should be enlarged, because the suck will not be as effective as in a normal baby. Other satisfactory methods are the use of eyedroppers or large syringes wilh rubber extension tubes connected to them. The tube is placed in the baby's mouth, and a small amount of solution is injected. These methods of feeding, while adequate for sustenance, require more time and care. Because the child will swallow a considerable amount of air when these feeding methods are used, the child is not usually fed while recumbent., and more frequent burping is necessary.

Ear Problems

Children afflicted with a cleft of the soft palate are predisposed to middle ear infections. The reason for this becomes clear on review of the anatomy of the soft palate musculature, The levator veli palatini and tensor vclj palatini, which are normally inserted into the same muscles on the opposite side, are left unattached when the soft palate is cleft. These muscles have their origins either directly on or near the auditory tube. These muscles allow opening of the ostium of this tube into the nasopharynx. This action is demonstrated when middle ear pressures are equalized by swallowing during changes in atmospheric pressure, as when ascending or descending in an airplane.

When this function is disrupted, the middle ear is essentially a closed space, without a drainage mechanism.

Serous fluid may then accumulate and result in serous otitis media. Should bacteria find their way from the nasopharynx into the middle ear, an infection can develop (i.e., suppurative otitis media). To make matters worse the auditory tube in infants is at an angle that does not promote dependent drainage. With age this angulation changes and allows more dependent drainage of the middle ear.

Children afflicted with cleft palate will frequently need to have their middle ear "vented." The otorhinolaryngologist, who creates a hole through the inferior aspect of the tympanic membrane and inserts a small plastic tube, performs this procedure, which drains the ear to the outside instead of the nasopharynx (myringotomy).

Chronic serous otitis media is common among children with cleft palate, and multiple myringolomics arc frequently necessary. Chronic serous otitis media presents a serious threat to hearing. Because of the chronic inflammation in the middle ear, hearing impairments are common in patients with cleft palate. The type of hearing loss experienced by the patient with cleft palate is conductive, meaning that the neural pathway to the brain continues to function normally. The defect in these instances is simpiy that sound cannot reach the auditory sensory organ as efficiently as it should because of the chronic inflammatory changes in the middle ear. However, if the problem is not corrected, permanent damage to the auditory sensory nerves (i.e., sensory neural loss) can also result. This type of damage is irreparable. The range of hearing impairment found in individuals with cleft palates is vast. The loss can be great enough so that normal-sounding speech is heard at less than one half of expected volume. In addition, certain sounds of speech (called *phonemes),* such as the *s, sh,* and *t* sounds, may be heard poorly. Audiograms are useful tools and arc performed repeatedly on patients with cleft palates, to monilor hearing ability and performance.

Speech Difficulties

Four speech problems are usually created by cleft lip and palate deformity. Retardation of consonant sounds (i.e.,*#, b, i, d, k, g)* is the most common finding. Because consonant sounds are necessary for the development of early vocabulary, much language activity is omitted. As a result, good sound discrimination is lacking by the time the palate is closed. I lypernasality is usual in the patient with a cleft of the soft palate and may remain after surgical correction. Dental malformation, malocdusiun, and abnormal tongue placement may develop before the palate is closed and thus produce an articulation problem. Hearing problems contribute significantly to the many speech disorders common in patients with oral clefts.

In the normal individual, speech is created by the following scheme. Air is allowed to escape from the lungs, pass through the vocal cords, and enter the oral cavity. The position of the tongue., lips, lower jaw, and soft palate working together in a highly coordinated fashion results in the sounds of speech being produced. If the vocal cords are set into vibration while the airstream is passing between, then voice is superimposed on the speech sounds that result from the relationships of the oral structures.

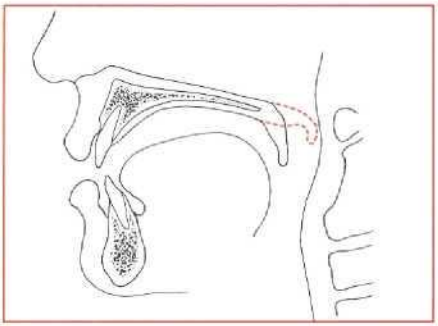


FIG- 27-7 Upward and backward movement of soft palate during normal speech. Its contact with posterior pharyngeal wall is shown.

The soft palate is raised during speech production, preventing air from escaping through the nose.

For clear speech it is necessary tor the individual to have complete control of the passage of air from the oropharynx to the nasopharynx. The hard palate provides the partition between the nasal and oral cavities. The soft palate functions as an important valve to control the distribution of escaping air between the oropharynx and nasopharynx (Fig. 27-7). This is called the *veiopharyngeui mechanism {vela* means *soft palate).* As the name implies, Its two main components are (1) the soft palate and (2) the pharyngeal walls. When passive the soft palate hangs downward toward the tongue, but during speech the muscles of the soft palate elevate it and draw it toward the posterior pharyngeal wall, which is what happens to the normal individual's soft palate when he or she is asked to say *"ah."* In normal speech this action takes place rapidly and with an unbelievable complexity, so that the valving mechanism tan allow large amounts of air to escape into the nasopharynx or can limit the escape to none.

In individuals whose soft palate is cleft, the velopharyngeal mechanism cannot function because of the discontinuity of the musculature from one side to the other. The soft palate thus cannot elevate to make contact with the pharyngeal wall. The result of this constant escape of air into the nasal cavity is *hypeniasal speech.*

Individuals with cleft palate develop compensatory velopharyngeal, tongue, and nasal mechanisms in an attempt to produce intelligible speech. The posterior and lateral pharyngeal walls obtain great mobility and attempt to narrow the passageway between the oropharynx and nasopharynx during speech. A muscular bulge of the pharyngcal wall actually develops during attempts at closure of the passageway in some individuals with cleft palate and is known as *Passavant's ridge* or *bar.* Individuals with cleft palates develop compensatory tongue postures and positions during speech to help valve the air coming from the[1] larynx into the phyryngeal areas. Similarly, flic superficial muscles around the nose involved in

facial expression are recruited to help limit the amount of air escaping from the nasal cavity. In this instance the valving is at the other end of the nasal cavity from the velopharyngeal mechanism. However, in an uncorrected cleft of the soft palate, it is literally impossible for compensatory mechanisms to produce a satisfactory velopharyngeal mechanism. Unfortunately in surgically corrected soft palates, velopharyngeal competence is not always achieved with one operation, and secondary procedures are frequently necessary.

Speech pathologists are well versed in assisting children with deft deformities to develop normal articulation skills. The earlier in life speech training is started in patients with cleft deformities, the better the eventual outcome. The patient may need to undergo speech counseling for several years to produce acceptable speech.

When hearing problems are also present, the speech problems arc compounded. Hearing loss at an early age is especially detrimental to the development of normal speech skills. The child who is unable to hear is unable to imitate normal speech. Thus the parents must be cognizant of their child's development and ensure thai regular visits to the pediatrician are undertaken.

Associated Anomalies

Although the child with an oral cleft is 20 times more likely to have another congenital anomaly than a normal child, no correlation is evident with specific anatomic zones of additional anomaly involvement^ Of those children who have associated anomalies, 38% have isolated deft palate and 21% have cleft lip, with or without cleft palate. In the overall deft-afflicted population, approximately 30% have other anomalies in addition to the facial cleft, ranging from clubfootto neurologic disturbances. Of the every]] cleft-afflicted population, 10% have congenital heart disease, and 10% have some degree of mental retardation. Thus the child with a facial cleft may require additional care beyond the scope of the cleft team.

TREATMENT OF CLEFT LIP AND PALATE

The aim of treatment of cleft lip and palate is to correct the cleft and associated problems surgically and thus hide the anomaly so that patients can lead normal lives. This correction involves surgically producing a face that does not attract attention., a vocal apparatus that permits intelligible speech, and a dentition that allows optimal function and esthetics. Operations begin early in life and may continue for several years. In view of the gross distortion of tissues surrounding the cleft, it is amazing that success is ever achieved. However, with modern anesthetic techniques, excellent pediatric care centers, and surgeons who have had a wealth of experience because of the frequency of the cleft deformity, acceptable results are commonplace.

Timing of Surgical Repair

The timing of the surgical repair has been and remains one of the most debated issues among surgeons, speech pathologists, audlologisls, and orthodontists, It is tempting to correct all of the defects as soon as the baby is able to withstand the surgical procedure. The parents of a child born with a facial cleft would certainly desire this mode of treatment, eliminating all of the baby's clefts as early in life as possible. Indeed the cleft lip is usually corrected as early as possible. Most surgeons adhere to the proven "rule of 10" as determining when an otherwise healthy baby is fit for surgery (i.e., 10 weeks of age, 10 lb in body weight, and at least 10 g of hemoglobin per dccilitci of blood). However, because surgical correction of the cleft is an elective procedure, if any other medical condition jeopardizes the health of the baby, the cleft surgery is postponed until medical risks are minimal.

Unfortunately each possible advantage for closing a palatal cleft early in life has several possible disadvantages for the individual later in life. The six advantages for early closure of palatal defects are (1) better palatal and pharyngeal muscle development once repaired, (2) ease of feeding, (3) better development of phonation skills, (4J better auditory tube function, (5) better hygiene when the oral and nasal partition is competent, and (6) improved psychologic state for parents and baby. The disadvantages of closing palatal clefts early in life are also several: The two most important are (1) surgical correction is more difficult in younger children with small structures, and (2) scar formation resulting from the surgery causes maxillary growth restriction.

Although different cleft teams time the surgical repaiT differently, a widely accepted principle is compromise. The lip is corrected as early as is medically possible, The soft palatal cleft is closed between 8 and 18 months of age, depending upon a host of factors. Closure of the lip as early as possible is advantageous, because it performs a favorable "molding" action on the distorted alveolus. It also assists the child in feeding and is of psychologic benefit. The palatal cleft is closed next, to produce a functional velopharyngeal mechanism when or before speech skills are developing. The hard palatal cleft is occasionally not repaired at the time of soft palate repair, especially if the cleft is wide. In such cases, the hard palate cleft is left open as long as passible so that maxillary growth will proceed as unimpeded as possible. Closure of the hard palatal cleft can be postponed at least until all of the deciduous dentition has erupted. This postponement facilitates the use of orthodontic appliances and allows more maxillary growth to occur before scarring from the surgery is induced. Because a significant portion of maxillary growth has already occurred by ages 4 to 5, closure of the hard palate at this time is usually performed before the child's enrollment in school. Removable palatal obturators can be fitted and worn in the meantime to partition the oral and nasal cavities.

The largest problem in evaluation of treatment regimens is the fact that the final results of surgical repair of clefts can only be fudged conclusively when the individual's growth is complete. A surgical method used today cannot be put to careful scrutiny for 10 to 20 years, which, unfortunately, may allow many individuals with cleft deformities to be treated with procedures that may later be discarded, when follow-up examinations and studies show unsatisfactory or poor effects.

Cheilorrhaphy

Cheilorrhaphy is the surgical correction of the cleft lip deformity; this term is derived from *ditilo,* lip, and *rhaphy,* junction by a seam or suture. It is usually the earliest operative procedure used to correct cleft deformities and is undertaken as soon as medically possible.

The cleft of the upper lip disrupts the important circumoral orbicularis oris musculature. The lack of continuity of this muscle allows the developing parts of the maxilla to grow in an uncoordinated manner so that the cleft in the alveolus is accentuated. At birth the alveolar process on the unaffected side may appear to protrude from the mouth. The lack of sphincteric muscle control from the orbicularis oris will cause a bilateral cleft lip to exhibit *a* premaxilla that protrudes from the base of the nose and produces an unsightly appearance. Thus restoration of this muscular sphincter with lip repair has a favorable effect on the developing alveolar segments.

Objectives. The objectives of cheilorrhaphy are twofold: (1) functional and (2) esthetic. The cheilorrhaphy should restore the functional arrangement of the orbicularis oris musculature to reestablish the normal function of the upper lip. If muscle continuity is not restored across the area of the cleft, an esthetically unpleasing depression will result when the lip is brought into function. The second objective of cheilorrhaphy is to produce a lip that displays normal anatomic structures, such as a vermilion tubercle, cupid's bow, and philtrum. The lip must be symmetric, well contoured, soft, and supple, and the scars must be inconspicuous. Another esthetic necessity is to correct (at least partially) the nasal deformity resulting from the cleft lip.

Despite the skill of the surgeon, these ideal objectives are rarely achieved. Hindrances are the poor quality of tissues in the cleft margins and the distortion of structures before surgical intervention. Several surgical techniques reproduce normal appearance immediately but do not maintain this appearance with growth. However, with careful selection of surgical technique, satisfactory results are obtainable.

Surgical techniques. As each cleft is unique, so must be the surgical procedure. Countless techniques can be used for cheilorrhaphy, each designed to elongate the cleft margins to facilitate closure (Figs. 27-8 and 27-9). In unilateral cases the unaffected side serves as a guide for lip length and symmetry, A key point in design is to break up lines of the scar so that with fibrosis and contracture, deformity of the lip will be minimized. In lips closed in a linear fashion, scar contracture causes a characteristic notching of the upper lip, Attention to reorienting and reuniting the musculature of the lip is of paramount importance if normal function is to be established.

Chcilorrhaphy procedures serve to restore symmetry not only to the lip but also to the nasal tip. With the cleft extending through the floor of the nose, the continuity of the nasal apparatus is disrupted. Without the bony foundation for the alar cartilage, a collapse of the lateral aspect of the nose occurs. When the lip is closed, it is necessary to advance this laterally displaced tissue toward the midline. Thus cheilorrhaphy is the first and one of the most important steps in correcting the nasal deformity so common in cleft patients.

Palatorrhaphy

Palatorrhaphy is usually performed in one operation, but occasionally it is performed in two. In two operations the soft palate closure (i.e., staphyloirhaphy) is usually performed first and. the hard palate closure (i.e., uranorrhaphy) is performed second.

Objectives, The primary purpose of the **cleft** palate repair is to create a mechanism capable of speech and deglutition without significantly interfering with subsequent maxillary growth. Thus creation of a competent velopharyngeal mechanism and partitioning of the nasal and oral cavities arc prerequisites to achieving these goals. The aim is to obtain a long and mobile soft palate capable of producing normal speech, lixtensivc stripping of soft tissues from bone will create more scar formation, which will adversely affect maxillary growth. The precarious nature of the problem indicates the complexity of the surgical procedures designed and the ages at which they are instituted.

Surgical techniques. Operative procedures for palatorrhaphy are as varied as techniques for cleft lip repair. Each cleft of the palate is unique. They vary in width, completeness, amount of hard and soft tissue available, and palatal length. Thus the surgical techniques used to close cleft palate deformities are extremely varied, not just from one surgeon to another but from one patient to the next.

Hard ***palate closure.*** The hard palate is closed with soft tissues only. Usually no effort is made to create an osseous partition between the nasal and oral cavities. The soft tissues extending around the cleft margin vary in quality. Some are atrophic and not particularly useful. Otheis appear healthy and readily lend themselves to dissection and suture integrity. In the most basic sense the soft tissues are incised along the cleft margin and dissected from the palatal shelves until approximation over the cleft defect is possible. This procedure frequently necessitates the use of lateral relaxing incisions close to the dentition (Fig. 27-10). The soft tissues are then sutured in a watertight manner over the cleft defect and allowed to heal. The areas of bone exposed by lateral relaxing incisions are allowed to heal by secondary intention. The superior aspect of the palatal flaps will also reepithelialize with respiratory epithelium, because this surface is now the lining of the nasal floor. When possible, it is advisable to obtain a two-layer closure Of the hard palatal cleft (Fig. 27-11 on page 637), which necessitates that the nasal mucosa from the floor, lateral wall, and septal areas of the nose be mobilized and sutured together before the oral closure.

When the vomer is long and attached to the palatal shelf opposite the cleft, a mucosa] flap can be raised from it and sutured to the palatal tissues on the cleft side (Fig. 27-12 on page 638). This procedure (i.e., vomer flap technique) requires little -stripping of palatal mucoperiosteum and produces minimal scar contraction. The denuded areas of vomer and the opposite sides of the flap where no epithelium is present will reepthelization. The vomer flap technique is useful in clefts that are not wide and where the vomer is readily available for use. it is a one-layer closure.

Soft palate closure. The closure of the soft palate is technically the most difficult of the operations yet discussed in the cleft-afflicted individual. Access is the

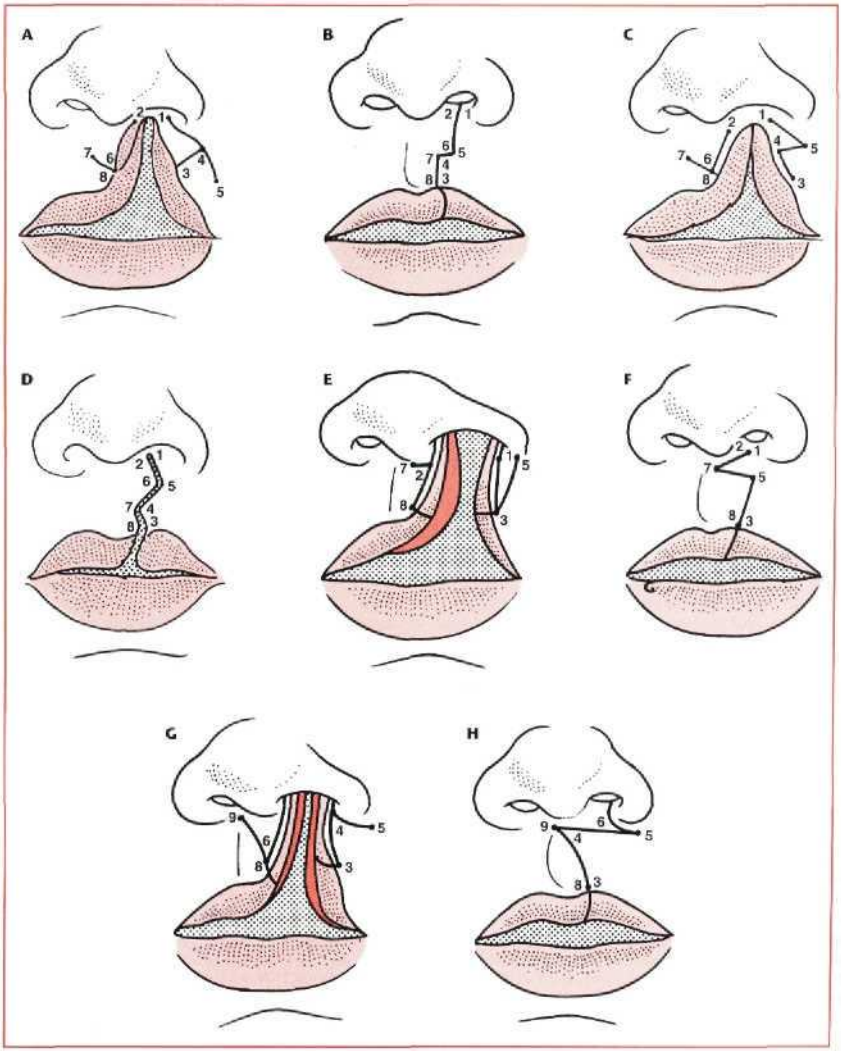


FIG. 27-8 Several cheilorrhophy techniques. A and B, *le.* Mesurier technique tor incomplete unilateral cleft. C and D.Tennison operation. E and F, Wynr operation. C and H, Millard operation (i.e.., rotation advancement technique).

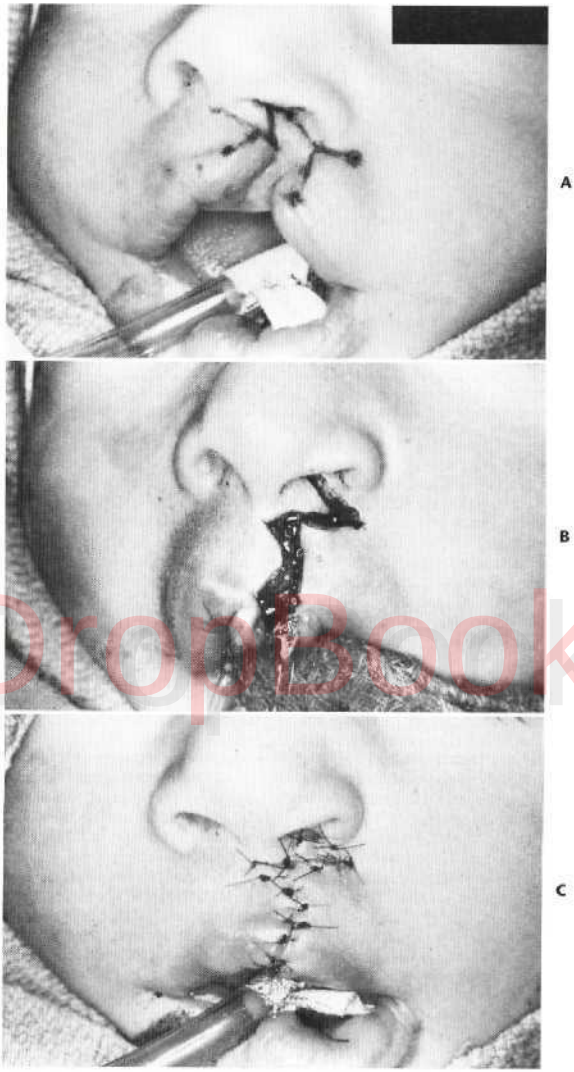


FIG. 27-9 The Millard cheilorrhaphy technique. A, Incisions outlined. B, Flaps rotated and advanced into position. C, Closure.

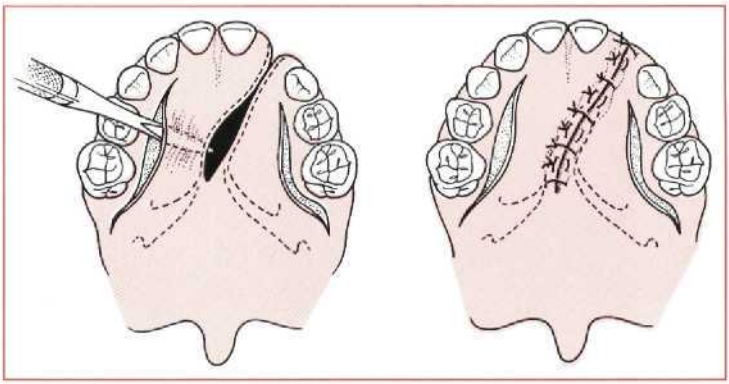


FIG. 27-10 Von Langenbeck operation for closure of hard palate iisfng lateral releasing incisions. This technique is one-layer closure—nasal (i.e., superior) aspect of palatal flaps will epithelialize, as will denuded areas of palatal bone.

largest problem, because the soft palate is toward the hack of Ihc oral cavity. The combination of difficulty with light, retraction, and the fact that the clinician can woik only from the oral side yet must correct both the oral and nasal sides of the soft palate lead to difficulties. In addition, tilt clinician may have to work with extremely thin, atrophic tissues yet produce a closure that will hold together under function while healing is progressing. To help accomplish this goal, the soft palate is always closed. in three layers and in the same order: (1) nasal mucosa, (2) muscle, and (3) oral mucosa (Fig. 27-13). The margins of the cleft are incised from the posterior end of the hard palate to *at least* the distal end of the uvula (some surgeons carry the incision and closure down the palatopharyngeal fold to elongate the soft palate). The nasal mucosa is then dissected free from the underlying musculature and sutured to the nasal mucosa of the opposite side. The muscular layer requires special care. The musculature of the deft soft palate is not inserted across to the opposite side but instead is inserted posteriorly and laterally along the margins of the hard palate. These muscular insertions must be released from their bony insertions and reapproximated to those of the other sides. Only then will the velopharyngeal mechanism have a chance to perform properly. If the quantity of muscular tissue is inadequate for approximation of the musculature in the mldllnc, the ptcrygoid hamular processes can be infractured, thus releasing the tensor palatini muscles toward the midline. This maneuver is frequently necessary, especially in wide clefts.

Occasionally, the soft palate is found to be short., and articulation with the pharyngeal wall is impossible. This situation is especially prevalent in incomplete palatal clefts—those of the soft palate only- In these cases the palate can be closed in a manner that not only brings the two lateral halves together in the midline but also gains palatal length (Fig. 27-14 on page 640). The so-called W-Y push-back procedure (Wardill) and U-shaped push-back procedure (Dorrance and Brawn) are commonly used. The mucoperiosteum of the hard palate is incised and elevated in a manner that allows the entire soft tissue elements of the hard and soft palate to extend posteriorly thus gaining palatal length.

Alveolar Cleft Grafts

The alveolar cleft defect is usually not corrected in the original surgical correction of either the cleft lip or the cleft palate (Fig. 27-15 on pages 641-642). As a result, the cleft-afflicted individual may have residual omnasal fistulae in this area, and the maxillary alveolus will not be continuous because of the cleft. Because of this, five problems commonly occur: (1) oral fluids escape into the nasal cavity, (2) nasal secretion drains into the oral cavity, (3) teeth erupt into the alveolar cleft, (4) the alveolar segments, collapse, and (5) if the cleft is large, speech is adversely affected,

Alveolar cleft bone grafts provide several advantages. First, they unite the alveolar segments and help prevent collapse and constriction of the dental arch, which is especially important if the maxilla has been orthodontically expanded. Second, alveolar cleft bone grafts provide bone support for teeth adjacent to the cleft and for those that will erupt into the area of the cleft. Frequently, the bone support on the distal aspect of the central incisor *is* thin, and the height of the bone support varies. These teeth may show slight mobility because of this lack of bone support. Increasing the amount of alveolar bone for this tooth will help ensure its periodontal maintenance. The canine tends to erupt into the cleft site and, with healthy bone placed into the cleft, will maintain good periodontal support during eruption and thereafter. The third benefit of alveolar cleft grafts is closure of the oronasal fistula, which will partition the oral and nasal

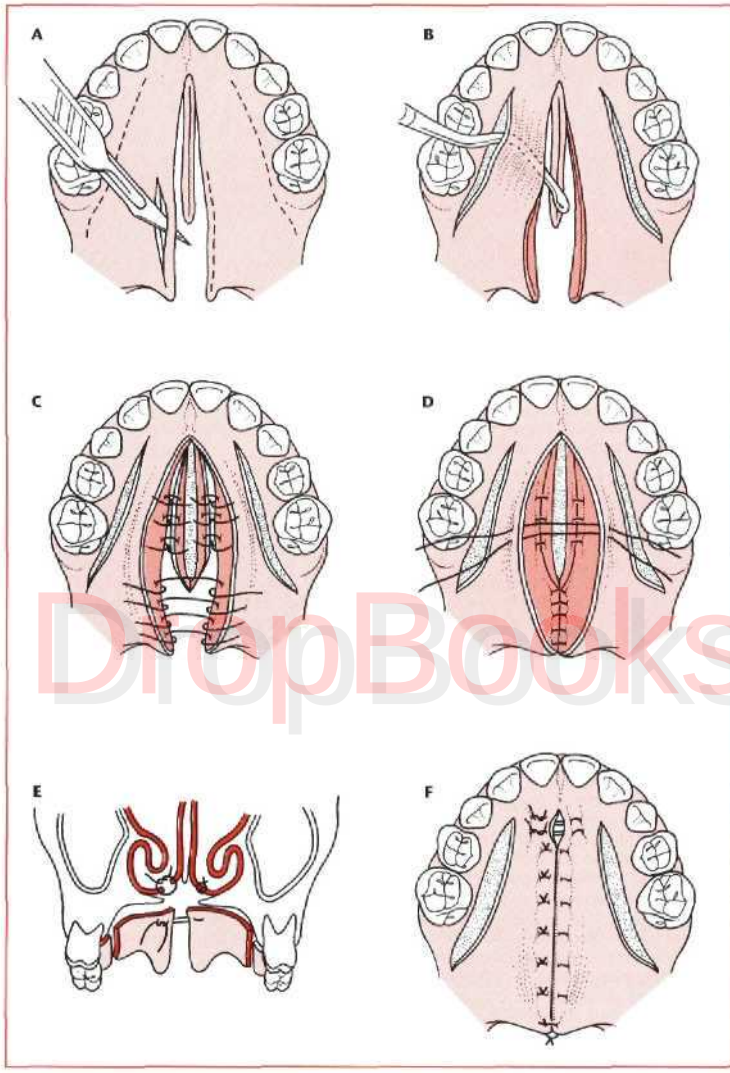


FIG. 27-11 Variation of von Langenbeck operation for concomitant hard and soft palate closure. It J5es three-layer closure for soft palate (i.e., nasal mucosa, muscle, oral mucosa) and two-layer closure for hard palate (i.e., flapi from vomer and nasal door to produce nasal closure, palatal flaps for oral closure). A, Removing mucosa from margin of cleft. B, Mucoperiosteal flaps on hard palate are developed; note lateral releasing incisions. C, Sutures placed into nasal mucosa after development of nasal flaps from vomer and nasal floor. Sutures are placed so that knots will be on nasal side. D, Nasal mucosa has been closed. E, Frontal section showing repair of nasal mucosa. F, Closure ol oral mucoperiosteum,

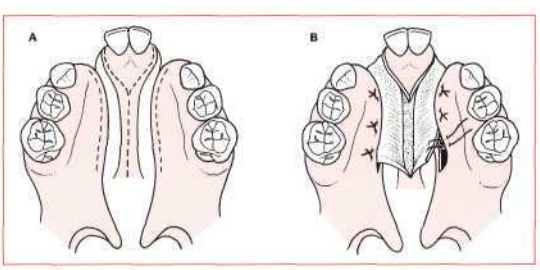


FIG. 27-12 Vomer flap technique for closure of hard palate cleft (bilateral in this case). A, Incisions through nasal mucosa on underside of nasal septum (i.e., vomer) and mucosa of cleft margins. B, Mucosa of nasal septum is dissected off nasal septum and inserted under palatal mucosa at margins Of cleft. This is one-layer nasal closure only. Connective tissue undersurface of nasal mucosa will epithelialize. This technique, because it does not require extensive elevation of palatal mucoperiosteum, producer less starring with attendant growth restriction.

cavities and prevent escape of fluids between them. Augmentation of the alveolar ridge in the area of the cleft is a fourth advantage, because it facilitates the use of dental prosthescs by creating a more suitable supporting base. A fifth benefit is the creation of a solid foundation for the lip and alar base of the nose. It has become evident that the alveolar cleft-grafting procedure itself creates a favorable change in the nasal structure, because the tissues at the base of the nose become supported after alveolar cleft grafting, whereas before the graft they had no solid osseous foundation. Therefore the alveolar graft should be performed before nasal revisions.

Timing of graft procedure. The alveolar cleft graft is usually performed when the patient is between ages 7 and 10. By this time a major portion of maxillary growth has occurred, and the alveolar cleft surgery should not adversely affect the future growth of the maxilla. It is important to have the graft in place before the eruption of the permanent canines into the cleft, thus ensuring their periodontal support. Ideally the grafting procedure is performed when one half to two thirds of the unerupted canine root has formed.

Orthodontic expansion of the arch before or after the procedure is equally effective; however, some surgeons prefer to expand before bone grafting so that access into the cleft area is facilitated at surgery.

Surgical procedure. Intact mucoperiosteal flaps on each side must cover bone grafts placed into the alveolar cleft. This means that flaps of nasal mucosa, palatal mucosa, and labial mucosa must all be developed and sutured in a tension-free, watertight manner to prevent infection of the graft. The soft tissue incisions for alveolar cleft grafts vary, but in each procedure these conditions are met (Fig. 27-16 on page 643).

The bone placed into the alveolar cleft is usually obUiincd trom I hi.[1] patient's iliutn or t: raniiim; however, some surgeons are using allogeneic bone (i.e., homologous bone from another individual). The grafts are made into a particulate consistency and are packed into the defect once the nasal and palatal mucosa have been closed. The labial mucosa is then closed over the bone graft. In time these grafts are replaced by new bone that is indistinguishable from the surrounding alveolar process (see Fig. 27-15). Orthodontic movement of teeth into the graft sites is possible, and eruption of teeth into them usually proceeds unimpeded.

Correction of Maxillomandibular Disharmonies

The individual with a cleft deformity will usually exhibit maxillary retrusion and a transverse maxillary constriction resulting from the cicatricial contraction of previous surgeries. In many instances the associated malocclusion is beyond the scope of orthodontic treatment alone. In these cases orthognathic surgery similar to the procedures outlined in Chapter 25 are indicated to correct the underlying skeletal malrclationships.

However, some differences exist in the technical aspects of maxillary surgery because of the other deformities and scarring that are present in the maxillas of cleft-afflicted individuals. In general, total maxillary osteotomies aTe necessary to advance and sometimes widen the maxilla. Closure of some of the space in the alveolar cleft area by bringing the alveolus of the cleft side anteriorly is also performed in several instances. These latter procedures necessitate the segmentation of the maxilla, which, because of the cleft's nature, usually already has occurred. The differences between the cleft-afflicted patient and a non-cleft-afflicted patient, however, arc the scar present across the palate and the decreased blood supply to the maxilla. Scarring from previous surgeries makes widening of the maxilla very difficult, and

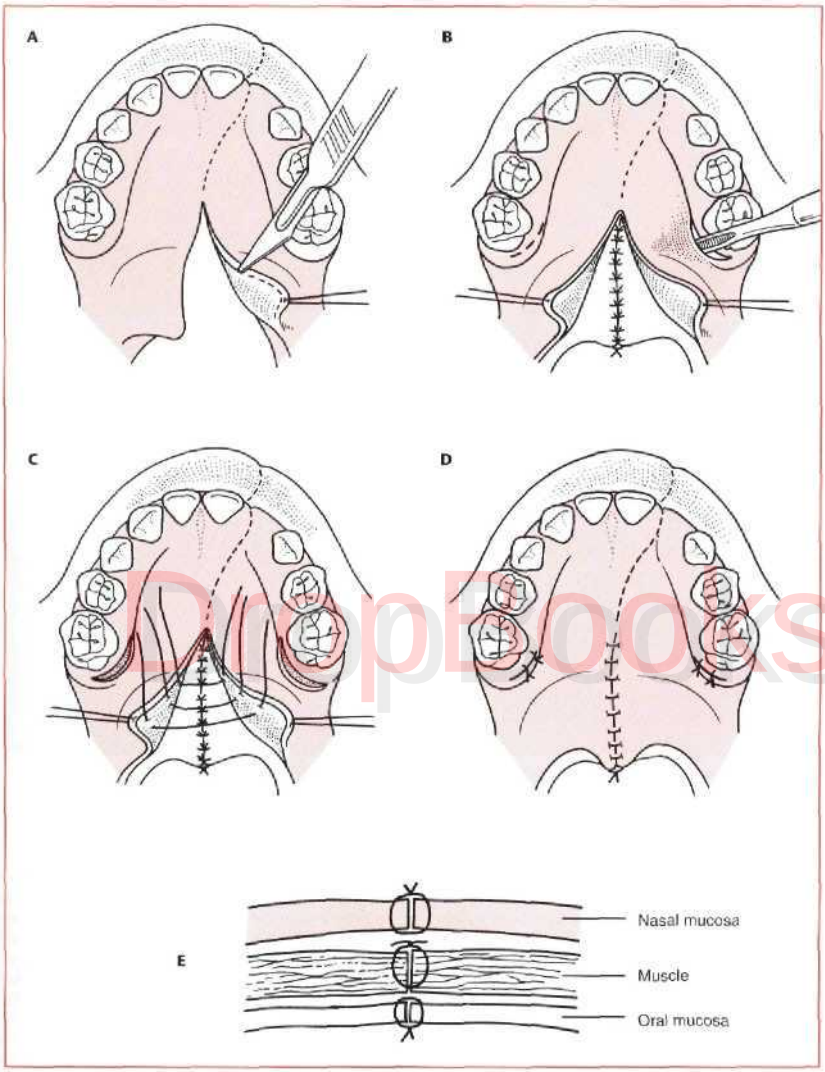


FIG. 27-13 Triple-layered soft palate closure. A, Excisfon of mucosa at cleft margin. B, Dissection of nasal mjcosa from soft palate to facilitate closure. Kasal mucosa is sutured together with knots tied or nasal (i.e., superior) surface. Note **small** incision made to insert instrument tor hamular process fracture. This maneuver refeases tensor veli palatini and facilitates approxímation in midline. C, Muscle *h* dissected from insertion into hard palate, and sutures are placed to approximate muscle in midline, D, Closure of oral mucosa is accomplished last. E, Layered closure of soft palate, *(from HaywofdfR:* Oral surgery, *Springfield, 111, 1976, Charles C Thomas,)*

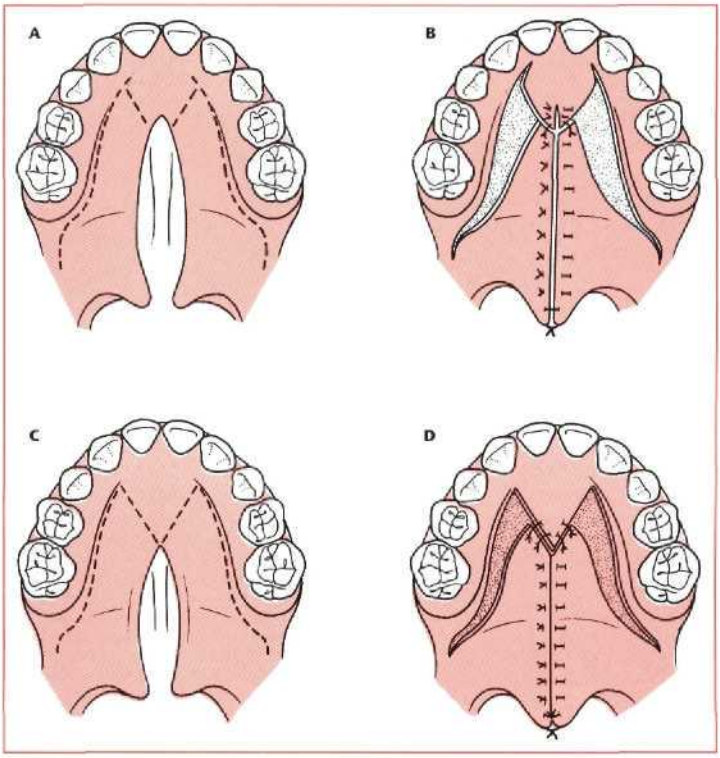


FIG. 27-14 The Wardill operations for palatal lengthening on closure, A and B, Four-flap operation for extensive cleft. C and D, Three-flap operation for shorter clefL Note amount of denuded palatal bone [eft after these operations.

frequently excision of some of this tissue is necessary. The clinician should try to be diligent and to maintain as much muooperlosteum to the maxilla as possible because of the poor blood supply that the deft maxilla receives. Care must also be taken not to create another oronasal fistula.

If the alveolar cleft had not been grafted previously, this can be done in the same operation. In bilateral clefts, however, the blood supply to the prolahial segment is very poor. It may be more prudent in these instances to perform the alveolar cleft grafts first and then perform a one-piece maxillary osteotomy after sufficient time has passed for revascularization of the prolabial segment,

One problem faced by the patient with a cleft palate when maxillary advancement procedures are planned is the effect this may have on the velopharyngeal mechanism. When the maxilla is brought forward, the. soft palate is also drawn forward. A patient's preoperativc marginal competence of the velopharyngeal mechanism may become incompetent in the postoperative period- it is very difficult to determine which patients will have this problem. Because of the possibility of this incompetence, however, secondary palatal or pharyngeal surgical procedures to increase velopharyngeal competence are discussed with the patient. These procedures can be performed later if necessary.

Secondary Surgical Procedures

Secondary surgical procedures are procedures performed after the initial repair of the cleft defects in an effort to improve speech or correct residual defects, The most commonly used technique to improve velopharyngeal competence secondarily is the pharyngeal flap procedure

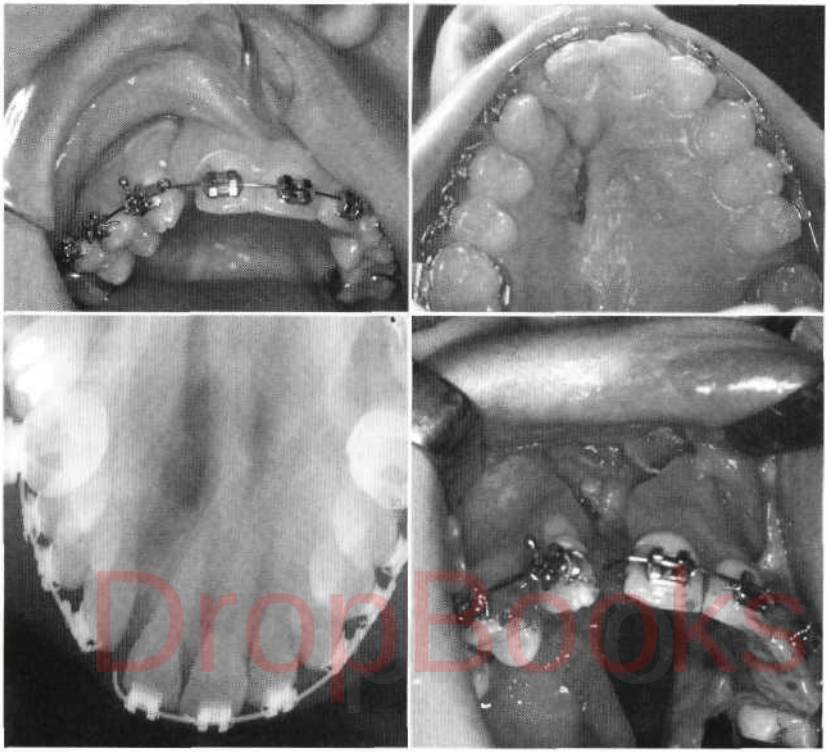


FIG. 27-15 Labial (A), palatal, (B) and radiographic (C) views of a patent unilateral atveolar cleft that extends posteriorly along the hard palate, D, Photograph shows surgical closure of the nasai rnucosa with inversion into the nasal cavity,

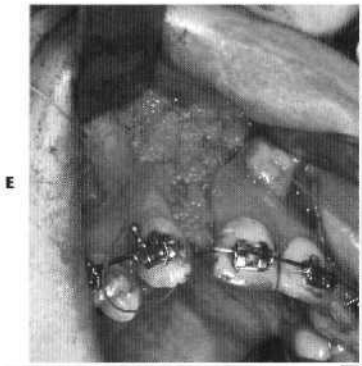
Continued

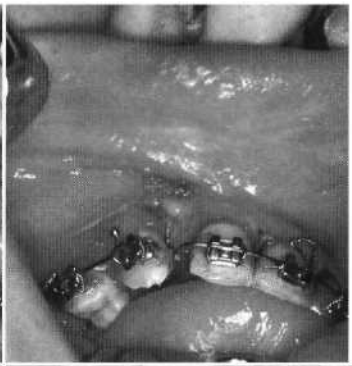
(Fig, 27-17). In this procedure a wide vertical strip of pharyngcal mucosa and musculature is raised from the posterior pharyngeal wall and inserted Into the superior aspect of the soft palate. These flaps are. most often based superiorly. The defect left in the posterior pharyngeal wall from elevation of the pharyngeal flap can be closed primarily or left to heal by secondary intention. Once inserted Into the soft palate, the pharynx and the soft palate arc joined, leaving two lateral ports as the opening between the oropharynx and nasopharynx, which reduces the airstream between the oropharynx and nasopharynx. The velopharyngeal mechanism then consist[1], of both raising the soft palate somewhat and medial constriction of the lateral pharyngeal walls.

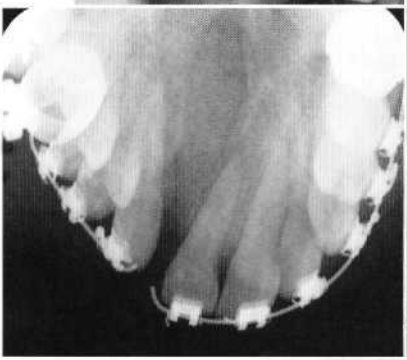
Another technique that has recently had a resurgence of interest because of new biocompatible material is the placement of an implant behind the posterior pharyngeal wall to bring it anteriorly (Fig. 27-18). Thus the soft palate has less distance to traverse to close off the nasopharynx. The major problems with this technique in the past have been migration of the implant and infection, which usually results in need for removal.

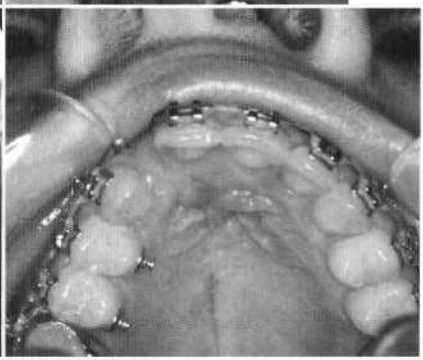
DENTAL NEEDS OF CLEFT-AFFLICTED INDIVIDUALS

Dentists will have cleft-afflicted patients in their practice because of the relatively large number of people so affected. These patients should not pose any great problems, because their dental needs do not differ dramatically from those of other individuals. However, because of the presence of the cleft, either corrected or uncorrccted, these individuals have a few special needs of which the dentist should be cognizant.









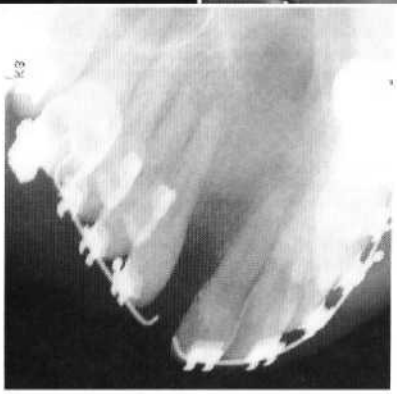


FIG. 27-15—cont'd E, Participate bone gratt is placed into the detect F, Closure of the palatal and labiaJ mucosa over the bone graft, G, Radiographic result is demonstrated 3 days after surgery. H, Three months later the soft tissues have healed. I, Radiograph shows consolidation of the bone grsfr.

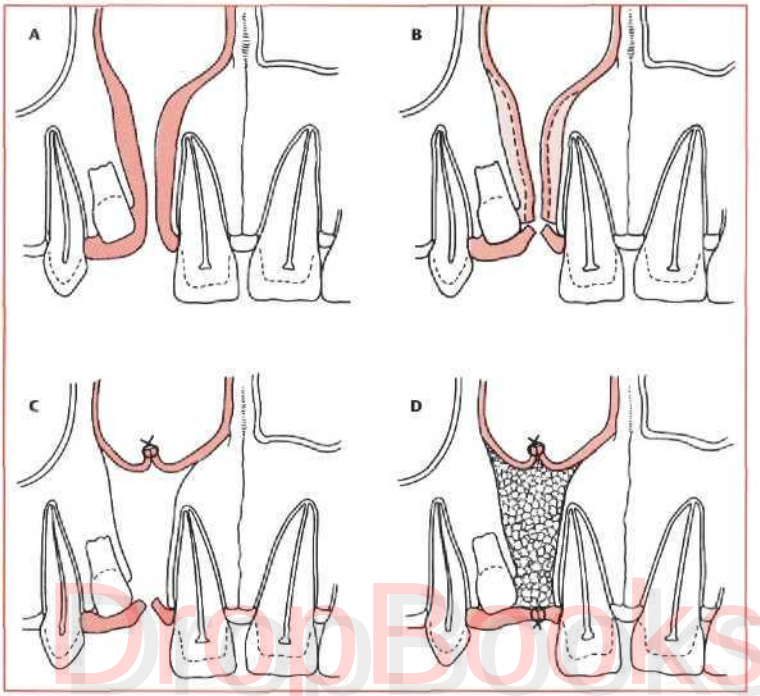


FIG. 27-16 Technique for alveolar cleft bone grafting. A, Preoperative defect viewed from labial aspect. Fistula extends, into nasal cavity. B, Incision divides mucosa fistula, which allows development of nasal and oral flaps. C, Mucosal flap developed from lining of fistula is turned inward, up into nasal cavity, and sutured in watertight manner D, Bone graft material is packed into cleft, and oral mucosa is closed in watertight manner.

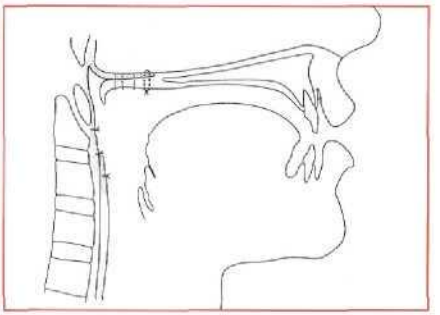


FIG. 27-17 Superiorly based pharyngeal flap. Flap is sutured to superior aspect of soft paiate, thus partially partitioning oral and nasal cavities from one another. Only nasal airway remaining after this operation is two lateral openings on each side of flap.

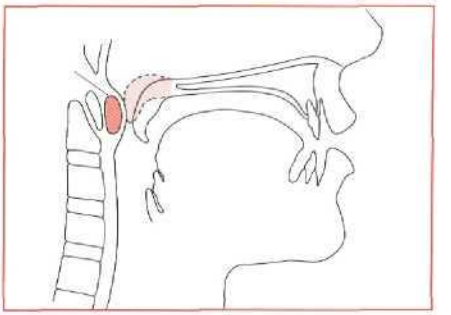


FIG. 27-18 Posterior pharyngeai wall implant. This makes distance between soft palate and pharyngeal wall smaller so that velopharyngeal closure is facilitated.

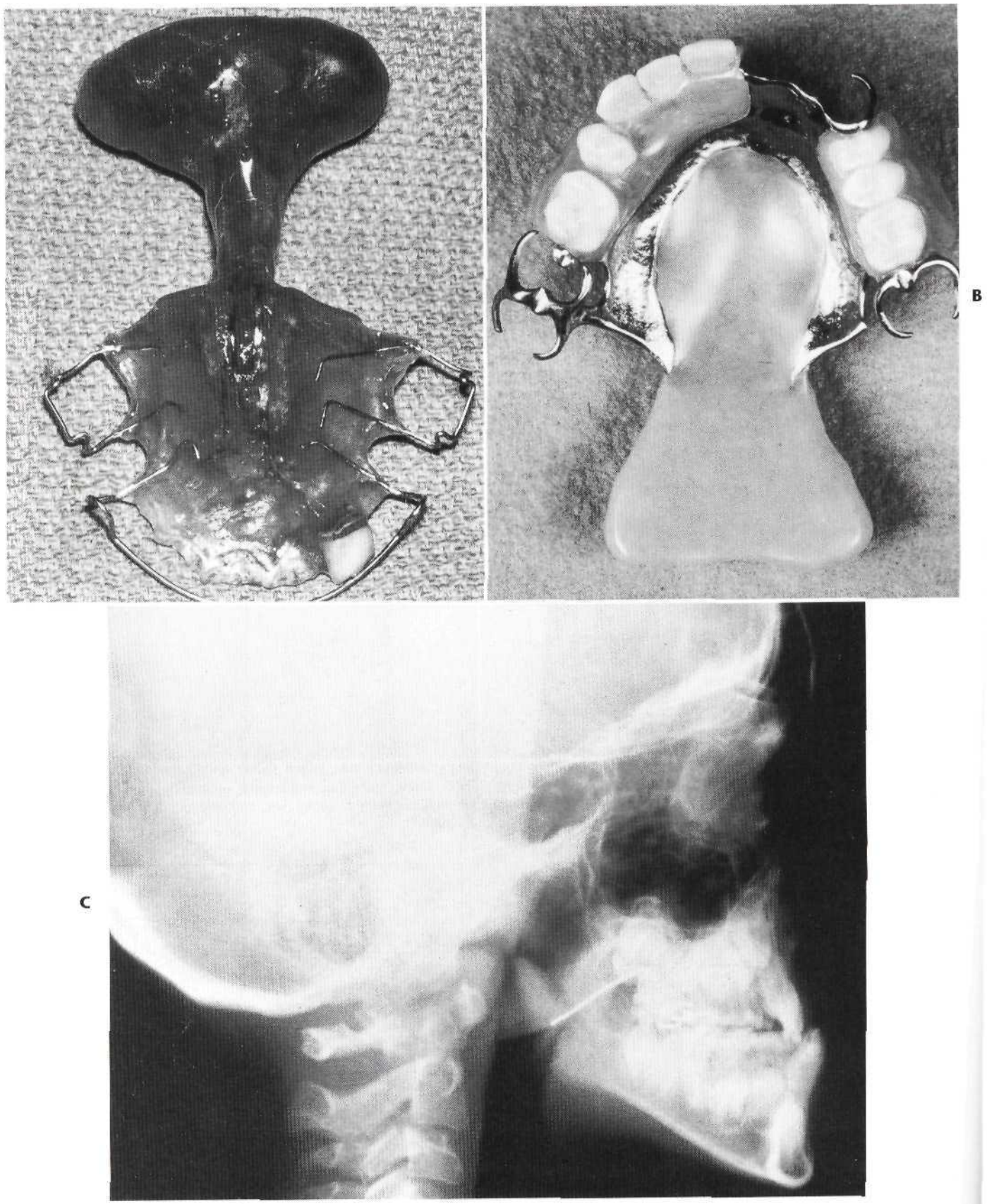


FIG. 27-19 Prosthetic speech aid appliances. A and B, Appliances designed both to lift solt palate and to obturate oral and nasal cavities. C, Lateral cephalogram with appliance in place. Note acrylic bulb positioned between soft **palate** and posterior **pharyngeal** wall.

Because of the interdisciplinary approach that cleft-afflicted patients require, it behooves the dentist to be aware of the overall treatment plan formulated by the deft team for Ihe patient's management. Awareness of this plan precludes the performance of any irreversible or costly procedures on teeth that may be charted for extraction in the future. For instance, placing a bridge to replace a congenially missing lateral incisor before alveolar bone grafting and orthodontic therapy is unwise. Similarly, extracting supernumerary teeth lhat may be temporarily retained to maintain alveolar bone support is also disadvantageous. All fixed bridgework should be delayed until after the orthodontic, orthognathic, and alveolar grafting procedures have been completed. Only then will the dentist be able to determine accurately the exact space and ridge form available for pontics. Furthermore, until the two halves of the maxillary arch have been united with bone grafts, the halves will move independently and bridgework spanning the cleft margin may become loose. Therefore the dentist must communicate freely with the other professionals who are managing the patient's other cleft problems, and coordination of services is of paramount importance.

Teeth adjacent to the cleft margins not only may be malformed or absent but also may have poor periodonlal support because of lack of bone and their position in the deft margin. This situation predisposes them to periodontiUs and early loss if nol kept in an optimal state of health, because teeth are Frequently malaligned and rotated, oral hygienic measures may be more difficult; these individuals may need more frequent prophylaxis and special oral hygienic instructions with careful reinforcement. Otherwise, rampant caries with premature loss may occur. This is a special tragedy in the cleft-afflicted individual, because he or she may have fewer teeth to serve vital functions (e.g., retaining orthodontic, orthopedic, or speech appliances).

Prosthetic Speech Aid Appliances

Prosthetic care for the cleft patient may he necessary for two reasons: First, teeth that are so frequently missing in the cleft-afflicted patient should be replaced. Second, in patients who have failed to obtain velopharyngcal competence with surgical corrections, a speech aid appliance can be made by the dentist to decrease hypernasal speech. A speech aid appliance is an acrylic bulb attached to a tooth-borne appliance in the maxilla (Fig. 27-19). The btilb is fitted to project onto the undersurface of the soft palate and lifts the soft paiate superiorly. If this bulb does not £ive adequate function, another projection of acrylic (i.e., bulb obturator) can be placed lo extend to the posterior aspect of the palate. This narrows the pharyngeal isthmus, and the size can be adjusted lor maximal effectiveness. The poslerior pharyngeal wail then will contact this bulb in function. In many instances the size of the bulb can he reduced as the pharyngeal musculature becomes more active. This type of appliance is used in two instances: (l) before a pharyngeal flap procedure to develop muscle action or (2) if the secondary surgical procedures are not successful in producing velopharyngeal competence. The speech aid appliance is also useful concomitantly to hold prosthetic dental replacements, to cover hard palate defects, and to support deficient upper lips by a flange extending into the labial stilcus. Obviously the maintenance of the residual dentition in an optimal state is prerequisite for successful speech aid appliance therapy.

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